[ASN Abstract: 2500-character + spaces limit; count includes title and author names; max 1 table counted as 50 characters per row and up to 2 Figures with max 560 characters each]

Current Count: 2493/2500 characters + spaces [131+136+1691+210+63+245=2493]

Abstract Title: [131]
Preliminary Findings From the Phase 2 EPPIK Study of Sparsentan in Pediatric Patients With Selected Proteinuric Glomerular Diseases

Author names: [136]
Howard Trachtman, Rosanna Coppo, Ping He, Kenneth V Lieberman, Alex Mercer, Michelle Rheault, Moin A Saleem, Teresa Shafer, Radko Komers

Author Affiliations: [not counted]
Howard Trachtman: University of Michigan, Ann Arbor, Michigan, USA
Rosanna Coppo: Fondazione Ricerca Molinette, Regina Margherita Hospital, Turin, Italy
Ping He, Radko Komers, Teresa Shafer: Travere Therapeutics Inc, San Diego, California, USA
Kenneth V Lieberman: Joseph M. Sanzari Children's Hospital of the Hackensack Meridian Health
Alex Mercer: JAMCO Pharma Consulting, Stockholm, Sweden
Michelle Rheault: University of Minnesota Medical School, Minneapolis, MN, USA
Moin A Saleem: University of Bristol & Bristol Royal Hospital for Children, Bristol, UK
Network, Hackensack, New Jersey, USA

Background: [593]
Sparsentan (SPAR) is a novel, non-immunosuppressive, single molecule Dual Endothelin Angiotensin Receptor Antagonist (DEARA) approved by the US FDA for the treatment of adults with IgA nephropathy (IgAN) at risk of rapid disease progression and is being investigated for focal segmental glomerulosclerosis (FSGS). The ongoing Phase 2 EPPIK study is examining the safety and long-term antiproteinuric and nephroprotective potential of SPAR in pediatric patients with FSGS, minimal change disease (MCD), IgAN, IgA vasculitis (IgAV), and Alport syndrome (AS). Here we report preliminary findings.

Methods: [602]
This open-label, single-arm, multicenter trial is evaluating the safety, efficacy, and pharmacokinetics of SPAR in ~30 patients aged 1-<18 years with FSGS and/or MCD (Population 1) and ~27 aged 2-<18 years with IgAN, IgAV, or AS (Population 2) over 108 weeks with a 4-week safety follow-up. SPAR is administered once daily in a liquid formulation with dose adjusted to body weight. Patients receiving RAASi undergo a 2-week washout prior to study medication start (baseline). Primary endpoints include safety and efficacy (change in urine protein/creatinine ratio [UP/C] from baseline over 108 weeks).

Results: [272]

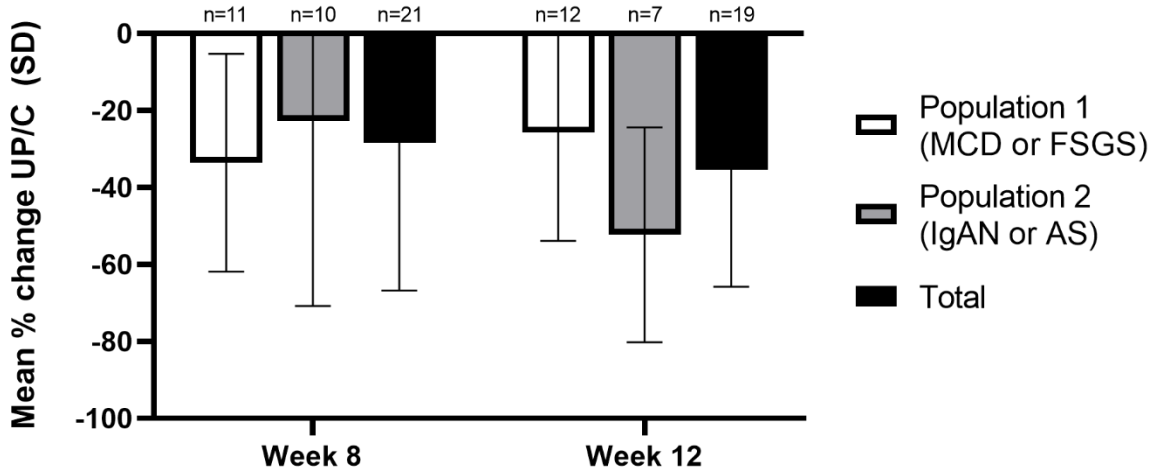
At data cutoff (4/5/23) 23 participants had received ≥1 dose of SPAR. Baseline characteristics are shown in **Table**. UP/C decreased over 12 weeks by 26% and 52% in Populations 1 and 2 and 35% overall (**Figure**). SPAR has been safe and generally well-tolerated.

Conclusion: [241]
SPAR treatment reduced proteinuria over the initial 12 weeks in pediatric patients with a range of proteinuric glomerular diseases. SPAR was safe and generally well-tolerated, consistent with findings from ongoing FSGS and adult IgAN trials.

Table: Baseline characteristics [210 characters; upload as a figure]

	Population 1 (n=13)	**Population 2 (n=10)**	**Total (N=23)**
MCD/FSGS, n (%)	8 (61.5) / 5 (38.5)	--	8 (34.8) / 5 (21.7)
IgAN/AS, n (%)	--	3 (30.0) / 7 (70.0)	3 (13.0) / 7 (30.4)
Age at screening, years, median (IQR)	8 (6, 13)	13 (12, 14)	12 (7, 14)
UP/CR, g/g, median (IQR)	3.0 (2.5, 5.7)	2.5 (2.1, 3.2)	2.8 (2.3, 5.0)
Nephrotic range proteinuria, UP/C ≥2 g/g, n (%)	12 (92.3)	8 (80.0)	20 (87.0)
eGFR, mL/min/1.73m^2, mean (SD)	106 (50)	87 (27)	98 (42)
Immunosuppression at baseline, n (%)	8 (61.5)	1 (10.0)	9 (39.1)

Figure: Mean percent change from baseline in UP/C with 12 weeks SPAR treatment [63 [title] + 245 [figure] characters; up to 2 figures allowed; character count determined by image height up to max of 560 characters]



SD bars are truncated at the upper end at 0.

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- Deadline for abstract submission: May 24, 2 p.m. EDT
- First author is presenter; author order set by submitter
- ALL authors must have an ASN User Login with current contact information and current financial and other relationships disclosure on file with ASN

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